A surgically treated case of Takayasu's arteritis complicated by aortic dissections localized in the ascending and abdominal aortae

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Because complication by aortic dissection is markedly rare in patients with Takayasu's arteritis, a limited number of reports have been published regarding surgically treated cases of Takayasu's arteritis that is complicated by aortic dissection. When graft replacement of the ascending aorta and aortic arch and extra-anatomic bypass grafting were performed in a 72-year-old Japanese woman with Takayasu's arteritis, which was complicated by aortic dissections localized in the ascending and abdominal aortae, the postoperative course of this patient was satisfactory. It was considered that the media became friable in this patient because of the presence of Takayasu's arteritis and that hypertension that persisted for a long time caused the independent development of aortic dissections in the ascending and abdominal aortae. (J Vasc Surg 2000;31:1042-5.)

Nonspecific inflammation of the artery is induced by Takayasu's arteritis, which has a predilection for young women.[1-3] Presently, aortic dissection is not a rare disease because the diagnosis of aortic dissection has become easier as the result of recent advancements in various diagnostic techniques, such as computed tomography (CT) and magnetic resonance imaging. However, a limited number of reports have been published that describe surgically treated cases of Takayasu's arteritis that is complicated by aortic dissection.[4-7] We recently encountered a patient with Takayasu's arteritis that was complicated by aortic dissections localized in the ascending aorta and the abdominal aorta. When ascending and aortic arch replacement and extra-anatomic bypass grafting were performed, the postoperative course of this patient was satisfactory. Here, we report the course of this patient.

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CASE REPORT

The patient was a 72-year-old woman, in whom hypertension was detected approximately 30 years ago. Approximately 10 years ago, when the patient consulted a local physician because of the appearance of cervical discomfort, aortic dilatation was indicated by chest radiograph. Therefore further examination was performed in our hospital in 1983. As a result, CT demonstrated ascending aortic dissection that measured 6 cm in diameter, which was accompanied by marked calcification over the entire circumference of the ascending aorta and aortic dissection that was localized in the abdominal aorta. Moreover, descending aortic stenosis accompanied by calcification was also observed. Therefore the diagnosis of Takayasu's arteritis was established. Because there were no particular symptoms and surgical treatment was considered difficult, the patient was discharged; and antihypertensive therapy was performed at the outpatient clinic to follow the course of this patient. However, a recent chest radiograph demonstrated marked dilatation of the ascending aorta. Therefore the patient was readmitted to our hospital in February 1993. Symptoms on admission included elevated superior limb blood pressure (160/92 mm Hg), decreased ratio of inferior to superior limb blood pressures (0.56), and detection of vascular murmurs at the bilateral regions of the neck and the abdomen. The results of hematologic examinations (C-reactive protein, 0.3 mg/dL; erythrocyte sedimentation rate, 30 mm/60 min) demonstrated no inflammatory findings. Moreover, there were no abnormal findings on any other examinations. However, a chest radiograph demonstrated cardiac dilatation (cardiothoracic ratio, 60%) and calcification of the descending aorta. Body CT demonstrated ascending

aortic dilatation that measured approximately 10 cm in diameter together with a dissecting flap and marked calcification of the descending aorta (Fig 1, *A*). Aortic dissection localized in the abdominal aorta was also detected (Fig 1, *B*). Aortography demonstrated marked dilatations of the ascending aorta and cervical vessels and stenosis of the descending and abdominal aortae (Fig 2, *A*).

Severe calcification of the aorta was observed in this patient together with dilative and stenotic lesions. Therefore the patient was surgically treated on the basis of the diagnosis of Takayasu's arteritis complicated by aortic dissections localized in the ascending aorta and the abdominal aorta on July 1993. Cardiopulmonary bypass was initiated from both the right axillary and right femoral arteries. The patient was cooled down to 20°C. With the use of separated extracorporeal circulation, antegrade selective cerebral perfusion was started from both the right axillary and left common carotid arteries, and the ascending aorta was opened. Fig 3, *A*, shows the intraoperative findings. A dissecting flap was detected at the proximal portion of the ascending aorta, and there were old thrombi in the false lumen. Then, ascending and aortic arch replacement and reconstructive operation of two branches of the aortic arch were performed with the open distal method. Finally right axillofemoral arterial bypass grafting were performed after the patients was weaned from cardiopulmonary bypass (Fig 2, *B*). Fig 3, *B*, shows the pathohistologic findings of the ascending aortic wall obtained during the operation. Progressive intimal fibrosis and severe disruption and fibrous scar in the media were observed. In adventitia, focal lymphocytic infiltration and fibrosis involving the vasa vasorum and peripheral neurons were recognized. These findings were compatible with the scar stage of Takayasu's arteritis. The postoperative course of this patient was satisfactory after 49 months. Moreover, postoperative aortography did not demonstrate any problems in reconstructed regions (Fig 2, *B*), and ankle blood pressure index was risen to 1.0.

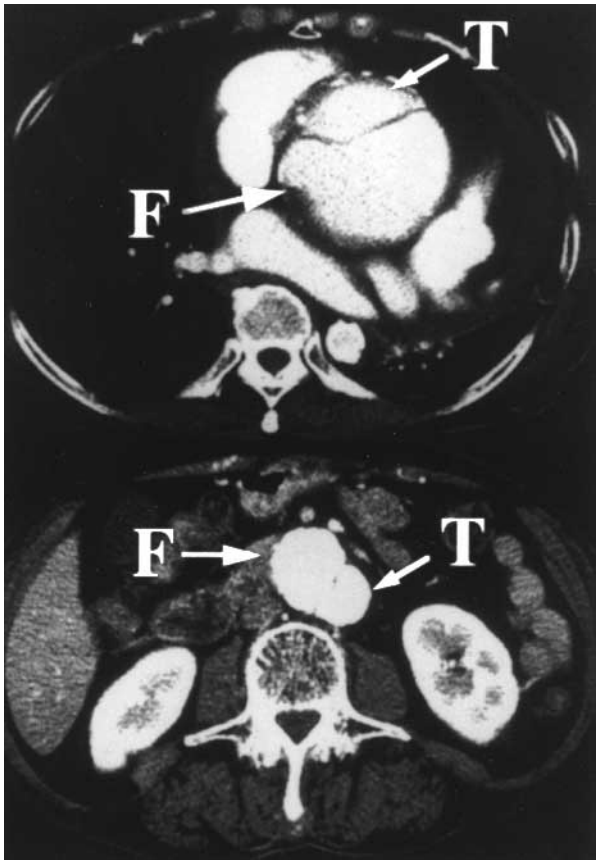

Fig 1. Preoperative CT findings. Aortic dissections were localized in the ascending aorta (Stanford type A) and the abdominal aorta, and CT showed marked calcification of the descending aorta. *T*, True lumen; *F*, false lumen.

DISCUSSION

Takayasu's arteritis is an inflammatory disease of unknown cause that is found mainly in young women and affects the aorta and its main branches.[1-3] It has a worldwide distribution but is more common in the Orient. Takayasu's arteritis was first reported in 1908 by Takayasu,[8] a Japanese ophthalmologist, as a peculiar condition that involves arteriovenous fistulas in the eyes. The diagnosis is usually initiated by a high index of clinical suspicion and requires a total arteriography to ascertain the pattern and distribution of arterial lesions. The diagnosis is confirmed whenever possible by examination of biopsy or autopsy specimens. In this patient, the diagnosis of Takayasu's arteritis was based not only on clinical and angiographic findings but also on pathologic findings in aortic wall specimens obtained during the operation. Pathologically, acute or active phase of Takayasu's arteritis was characterized by a granulomatous panarteritis with or without identifiable giant cells in the lymphoplasmacytic infiltrate. Healed lesions showed progressive intimal and adventitial fibrosis. Our patient also showed severe destruction and fibrosis in the media.

Because Takayasu's arteritis is accompanied by obstructive or dilative lesions, surgical treatment is indicated for this disease.[1-3] Between 1979 and 1998, 109 patients with Takayasu's arteritis underwent operation in our hospital. There were six men and 103 women. The age of these patients ranged from 15 to 76 years (mean, 48 years). Diagnoses that required an operation were obstruction of cervical vessels in 13 patients, coronary artery obstructive disease in 19 patients, pseudocoarctation of the aorta in 15 patients, obstruction of abdominal branches in 3 patients, dilatation of the aorta with aortic regurgitation in 44 patients, and thoracic aortic aneurysm in 41

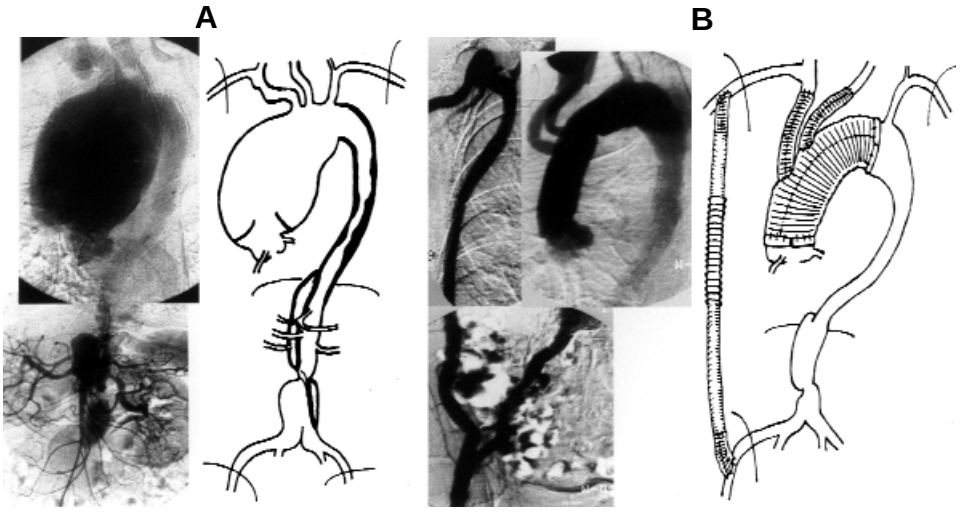

Fig 2. A, Preoperative angiography and anatomic scheme. B, Postoperative angiography and operative scheme. Ascending aorta and aortic arch replacement and extra-anatomic bypass grafting from right axillary artery to femoral artery were performed.

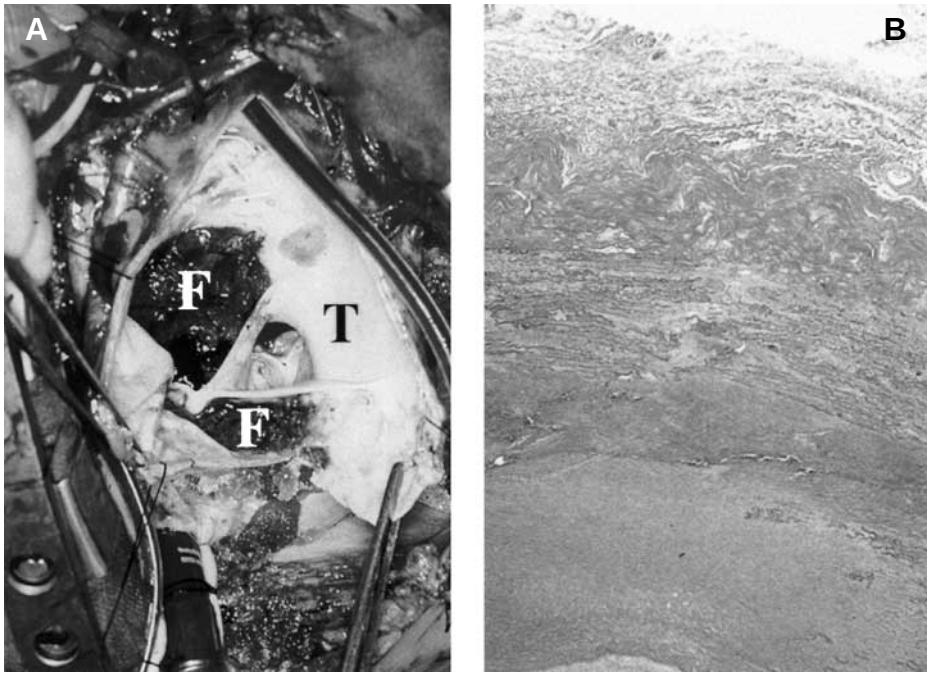

Fig 3. A, Intraoperative findings. A dissecting flap was detected at the proximal portion of the ascending aorta, and there were old thrombi in the false lumen. *T*, True lumen; *F*, false lumen. B, Pathologic findings of the specimen of the ascending aorta. Severe disruption and fibrosis were observed in the media, demonstrating the scar stage of Takayasu's arteritis (Elastica van Gieson stain).

patients. This patient had dilative lesion (thoracic aortic aneurysm) together with obstructive lesion (pseudocoarctation of the aorta). However, complication by aortic dissection is markedly rare, and only a limited number of surgically treated cases of Takayasu's arteritis, which was complicated by aortic dissection, were previously reported.[4-7] To date, 426 cases of aortic dissection were surgically treated in our

hospital. However, this was the first case of Takayasu's arteritis complicated by aortic dissection that was surgically treated in our hospital. It was previously reported that Takayasu's arteritis is more frequently complicated by aortic dissection localized in the descending or abdominal aorta (Stanford type B) than by aortic dissection localized in the ascending aorta (Stanford type A). Because aortic dissections in our patient were localized in the ascending and abdominal aortae, the respective aortic dissections were considered to have developed independently.

Although there are various causes of aortic dissection, hypertension, Marfan syndrome, and cystic medial necrosis are generally considered the major causes of aortic dissection.[9] In Takayasu's arteritis, the lesion extends to the respective layers of the arterial wall, but the media is most severely involved.[7] In addition, the pathologic findings of Takayasu's arteritis demonstrate disruption of medial elastic fibers. In our patient, marked stenotic lesion and calcification already occurred when Takayasu's arteritis was detected. Inflammatory findings were not observed at the time of operation, and the lesion was already in the course of involution. Moreover, hypertension of the superior limbs that persisted for a long time was induced by pseudocoarctation of the aorta because of abdominal aortic stenosis. Therefore it was considered that hypertension influenced the friable media, resulting in the development of aortic dissections in the ascending aorta and the region superior to the stenotic region of the abdominal aorta.

Although one aortic dissection was localized in the ascending aorta, dilatation of the aortic arch was also observed, and simultaneous graft replacement of the ascending aorta and aortic arch was performed.[10] Arterial blood flow through the cardiopulmonary bypass was obtained from both the axillary and femoral arteries because stenosis of the descending and abdominal aortae was observed. The cerebral circulation was preserved by selective cerebral perfusion during the reconstruction of ascending and aortic arch. Aortic dissection localized in the abdominal aorta remained untreated because the diameter of the abdominal aorta was not so large; the course of this patient was followed thereafter. Moreover, pseudocoarctation of the abdominal aorta was surgically treated with less invasive right axillofemoral arterial bypass grafting because of severe stenosis of the abdominal aorta, increased blood pressure difference between the superior and inferior limbs (70 mm Hg), and the occurrence of claudication. Wide lesions in the descending and abdominal aortae are frequently observed in Takayasu's arteritis. Therefore extra-anatomic bypass grafting is useful to treat such lesions because reconstructive operation can be performed in the region apart from these lesions. To treat pseudocoarctation of the aorta that is induced by Takayasu's arteritis, we previously performed ascending-abdominal aortic bypass grafting in four patients and axillary-iliac (femoral) arterial bypass grafting in 10 patients. As a result, the postoperative courses of these patients were all satisfactory.

REFERENCES

1. Kimoto S. The history and present status of aortic surgery in Japan particularly for aortitis syndrome. J Cardiovasc Surg 1979;20:107-26.
2. Takagi A, Tada Y, Sato O, Miyata T. Surgical treatment for Takayasu's arteritis: a long-term follow-up study. J Cardiovasc Surg 1989;30:553-8.
3. Lupi-Herrera E, Sanchez-Torres G, Marcushamer J, Mispireta J, Horwitz S, Vela JE. Takayasu's arteritis: clinical study of 107 cases. Am Heart J 1977;93:94-103.
4. Hachiya J, Tasaka A, Ishikawa T. Dissecting aneurysm in Takayasu's arteritis. Nippon Igaku Hoshasen Gakkai Zasshi 1973;33:578-85.
5. Lie JT. Segmental Takayasu (giant cell) aortitis with rupture and limited dissection. Hum Pathol 1987;18:1183-5.
6. Haga Y, Yoshizu H, Okuda E, Uriuda Y, Mitsumaru A, Tamai S, et al. Surgical reconstruction of an ascending aortic dissection in Takayasu's arteritis. Ann Thorac surg 1992;54:359-60.
7. Nakaji A, Ikezawa M, Kondo M, Kawakami M, Konno K, Takizawa Y, et al. A case of aortitis syndrome which presented coarctation of aorta due to intraluminal flap formation in the middle of the thoracic aorta. Kokyu To Junkan 1989;37:209-13.
8. Takayasu M. A case with unusual changes of the central vessel in the retina. Acta Soc Ophth Jap 1908;12:554-5.
9. Larson EW, Edwards WD. Risk factors for aortic dissection: a necropsy study of 161 cases. Am J Cardiol 1984;53:849-55.
10. Ando M, Nakajima N, Adachi S, Nakaya M, Kawashima Y. Simultaneous graft replacement of the ascending aorta and total aortic arch for type A aortic dissection. Ann Thorac Surg 1994;57:669-76.

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